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CORRESPONDENCE ANESTHESIA & CONCURRENT DISEASE

Anesthesia for Jervell and Lange Nielsen syndrome (JLNS)

Bilge Tuncer[1], Zeynep Şeyma Kadioğlu[2], Ezgi Erkiliç[3]

Authors affiliation:

1. Bilge Tuncer, Ankara Bilkent City Hospital, Ankara, Turkye; E-mail: bilgetuncer@yahoo.com
2. Zeynep Şeyma Kadioğlu, Ankara Bilkent City Hospital, Ankara, Turkye; E-mail: zynpsym@gmail.com
3. Ezgi Erkiliç, Ankara Bilkent City Hospital, Ankara, Turkye; E-mail: eerkilic72@yahoo.com

Correspondence: Ezgi Erkiliç; **E-mail:** eerkilic72@yahoo.com; **Mobile:** +90 5053663491

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To the editor,

Jervell and Lange Nielsen Syndrome (JLNS) is a rare autosomal recessive disorder. It is characterized by bilateral sensorineural hearing loss and a prolonged QTc interval, typically exceeding 500 msec. JLNS is a form of inherited long QT syndrome. Mutations in KCNQ1 and KCNE1 genes, responsible for cardiac K^+ channels, are associated with JLNS. These patients are at risk of developing polymorphic ventricular tachycardia (torsades de pointes) and ventricular fibrillation, which can lead to syncope and sudden death. It has been observed that surgical stress during the perioperative period and general anesthesia may trigger fatal arrhythmias.[1,2]

Beta-blockers are the first-line therapy for preventing syncope, cardiac arrest, and sudden death. If patients continue to experience symptoms despite beta-blocker treatment, implantable cardioverter-defibrillators (ICDs), pacemakers, or left cardiac sympathetic denervation may be considered.[3] Cochlear implantation is a successful treatment for hearing loss.[4]

A male patient, 4.5 y old and weighing 20 kg, with JLNS and a homozygous mutation in the KCNQ1 gene, was scheduled for right cochlear implantation. The patient was under cardiological follow-up for epicardial ICD implantation, ventricular tachycardia, and long QT and was taking propranolol. He had a history of adenoidectomy, bilateral ventilation tube insertion and removal, and ICD insertion. The preoperative physical examination was normal, and laboratory results were within normal ranges. The cardiac evaluation showed a heart rate of 103 beats per minute, indicating a paced rhythm. The QTc interval was 630 ms on the ECG. A preoperative adjustment was made to the dose of propranolol. On the day of the operation, the patient was sedated with midazolam 1 mg iv and taken to the operating room. In addition to the standard monitoring, a defibrillator and magnesium sulfate ($MgSO_4$) were prepared in case of an emergency. Anesthesia was induced using propofol 60 mg, fentanyl 20 µg, and rocuronium bromide 12 mg, along with lidocaine 20 mg. Intubation was completed without any issues. Anesthesia was maintained with infusions of 5 to 6 mg/kg/h of propofol and 0.1 µg/kg/min of remifentanil. The surgery lasted for 140 min and there were no instances of arrhythmia or hemodynamic deterioration. Postoperative pain management was achieved with intravenous morphine 1 mg and paracetamol 250 mg. The patient was extubated without any complications using sugammadex 40 mg.

Patients with JLNS require special attention to anesthetic concerns. Beta blockers should be continued during the perioperative period. Any form of sympathetic stimulation, such as anxiety, crying, or exposure to loud noise, can trigger arrhythmias. Inadequate anesthesia, bradycardia or tachycardia, hypertension, hypoxia, hypo- or hypercapnia, inadequate analgesia, and hypothermia may all cause sympathetic stimulation.[5,6] Therefore, we ensured that all of these factors were avoided and provided adequate analgesia for our patient. It is important to maintain normal serum electrolyte levels and prevent hypokalemia, hypomagnesemia, and hypocalcemia. Additionally, it is crucial to prevent high airway pressure. Beta-blockers should be readily available, as well as equipment for immediate

transcutaneous or transvenous pacing and defibrillation throughout the procedure.

It is recommended to avoid drugs that prolong the QT interval. Midazolam does not have an effect on the QT interval. Thiopental has been found to prolong the QT interval, while propofol has little to no effect on it. All volatile agents, except isoflurane, have the potential to prolong the QT interval. In these patients, vecuronium, atracurium, fentanyl, and morphine have been used without adverse effects. It should be noted that ondansetron may prolong the QT interval. The use of anticholinergic and anticholinesterase drugs as reversal agents, has been shown to prolong the QT interval.[2,3,5,6] Therefore, we used propofol, remifentanil, rocuronium bromide, and sugammadex instead. We also avoided using ondansetron.

Patients with LQTS present a challenge to anesthesiologists, and the situation can be even more difficult when dealing with undiagnosed patients. In conclusion, safe anesthesia management for children with JLNS requires perioperative continuation of beta blockers, avoidance of sympathetic stimulation, careful administration of anesthetics and other drugs to prevent QT prolongation, and the availability of magnesium sulfate and a defibrillator.

Conflict of interest

None declared by the authors.

REFERENCES

1. Schwartz PJ, Spazzolini C, Crotti L, Bathen J, Amlie JP, Timothy K, et al. The Jervell and Lange-Nielsen syndrome: Natural history, molecular basis, and clinical outcome. Circulation 2006;113:783-90.
2. Kies SJ, Pabelick CM, Hurley HA, White RD, Ackerman MJ. Anesthesia for patients with congenital long QT syndrome. Anesthesiology 2005;102:204-10.
3. Booker PD, Whyte SD, Ladusans EJ. Long QT syndrome and anesthesia. Br J Anaesth 2003;90:349-66.
4. Green JD, Schuh MJ, Maddern BR, Haymond J, Helffrich RA. Cochlear implantation in Jervell and Lange-Nielsen syndrome. Ann Otol Rhinol Laryngol Suppl. 2000;185:27-8.
5. Pabba K, Chakraborty RK. Jervell and Lange-Nielsen Syndrome. Internet: StatPearls Treasure Island (FL): StatPearls Publishing; 2023. PMID: 30725985. Bookshelf ID: NBK537300.
6. Niimi N, Yuki K, Zaleski K. Long QT Syndrome and Perioperative Torsades de Pointes: What the Anesthesiologist Should Know. J Cardiothorac Vasc Anesth 2022;36(1):286-302. doi: 10.1053/j.jvca.2020.12.011.

